ERUPTIVE XANTHOMAS AS A PRESENTING FEATURE OF DIABETES MELLITUS

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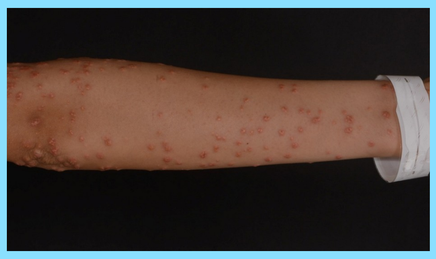
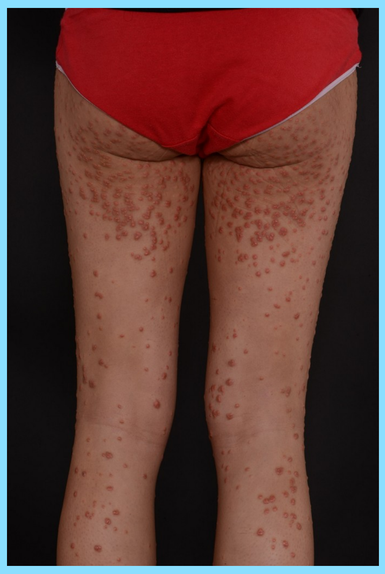
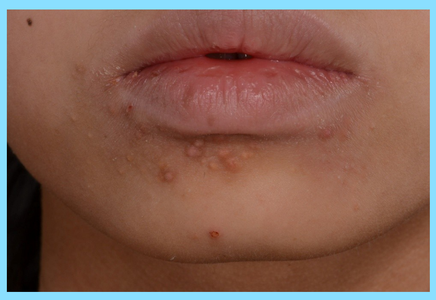
Presentation

A normally fit and well 14year old girl presented to Accident and Emergency with a six week history of widespread itchy, raised and painful rash. It had started over her neck and spread to her torso and limbs. She also complained of polydypsia and polyuria, although no weight loss. She denied any other symptoms including abdominal pain.

The family was from the Philippines and there was a strong history of Type 2 diabetes in her mother's family. She has two siblings who are both well.







The rash was diagnosed by the dermatology team as eruptive xanthomas.

Eruptive xanthomas are pink papules with creamy centres, often tender and itchy. They predominantly affect the trunk, buttocks and thighs. Histology reveals lipid laden histiocytes and lympho-neutrophilic infiltrate in the dermis. They resolve with improving lipid profile, usually over weeks.

Initial tests supported diagnoses of both hyperlipidaemia and new onset diabetes. ECG was normal.

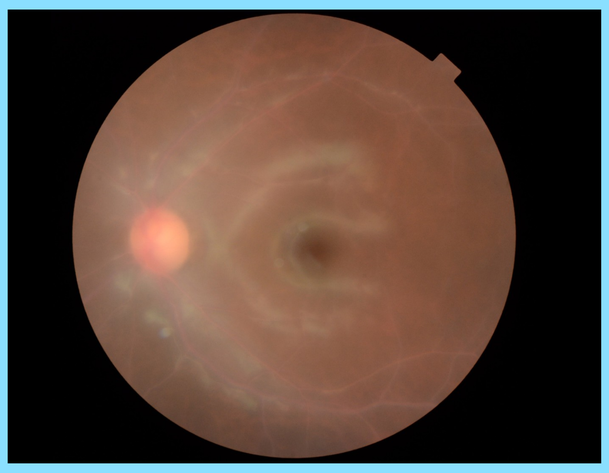
Blood gas
pH 7.36
CO_2 4.66
BE -5.3
Glucose 27
Lactate 0.8
Ketones 5.1

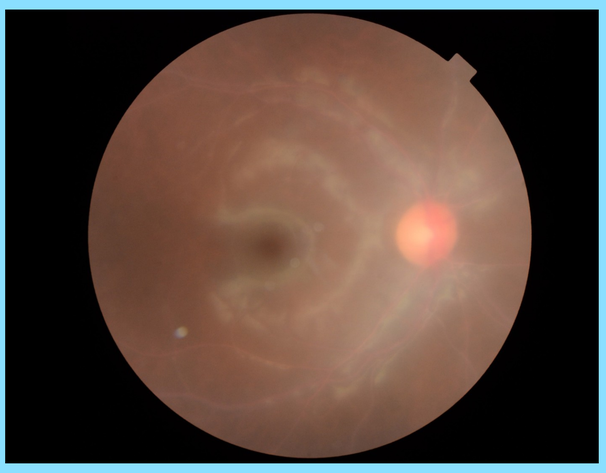
Admission blood Results

Cholesterol 34.2mmol (<5)
Triglycerides 189mmol (1.7)
HDL 0.8mmol (>1.2)
Amylase 85
HbA1c 17.7%
TSH 2.4, free T_4 19
C-peptide 0.35
Urate 216 (normall)

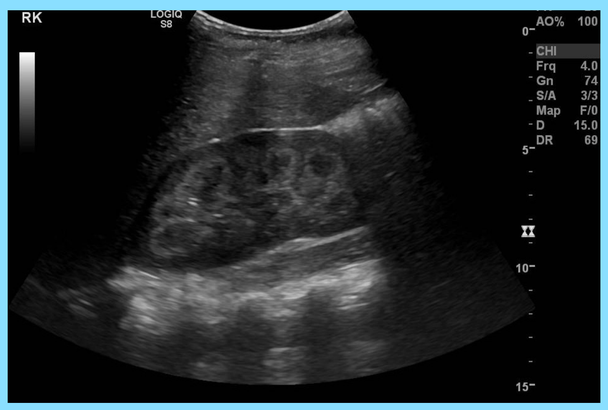
She had previously been for a routine eye check where she was told her eyes looked 'cloudy'. On ophthalmological examination she was diagnosed with lipaemia retinalis.

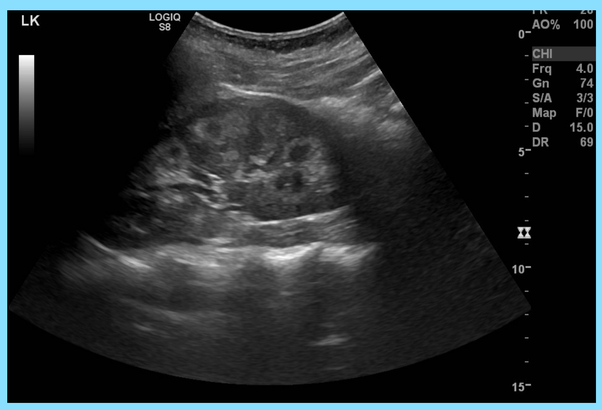
Lipaemia retinalis is a rare presentation occurring at triglyceride concentrations over 45mmol/l. The retinal vessels have a milky appearance, as seen below. This is due to visualisation of high levels of chylomicrons in the vessels and resolves as these lower.





Abdominal ultrasound scan showed xanthogranulomatous infiltration of both kidneys. This is demonstrated by distinctive bands of hyper-echogenicity around the pyramids seen in her pictures below.





Diabetic antibodies

Autoantibody screen negative
Anti-GAD <5U/ml
Anti-pancreatic islet cell Negative

Lipid electrophoresis – Type 5 hyperlipidaemia

Genetic analysis – no pathogenic mutations typical of Type 1 or 5 familial hyperlipidaemia

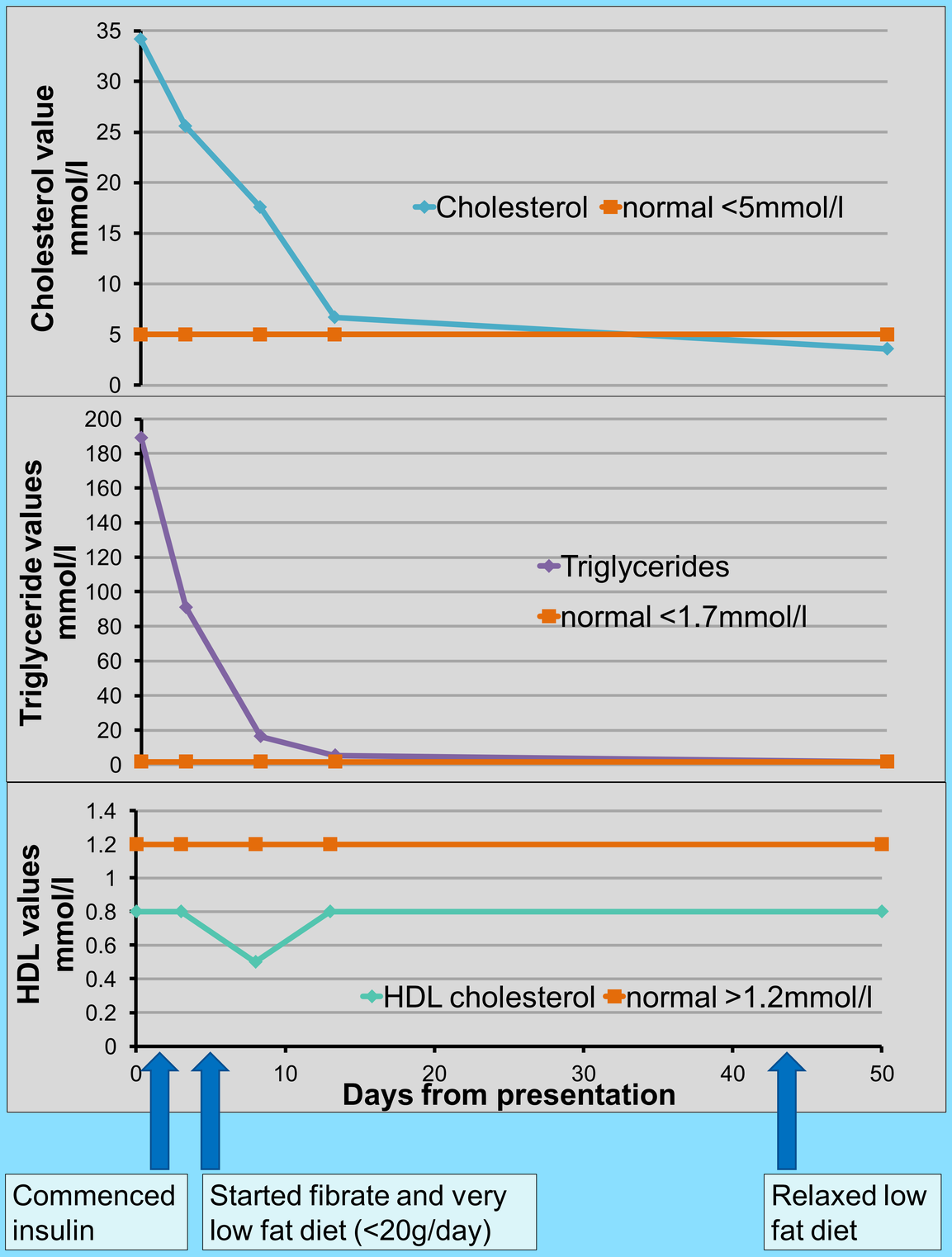
Given the extent of her hyperlipidaemia, further investigation was carried out to determine whether she had a primary familial hyperlipidaemia with associated insulin resistance, or whether her dyslipidaemia was secondary to uncontrolled diabetes.
Since there was no evidence of familial hyperlipidaemia we concluded the latter.

Management

Total 10 day admission for education and to achieve safe lipid profile

Diabetes:
Glucose control was initially erratic
Established on Levemir and Novorapid basal bolus regime
Discharged on 0.8units/kg/day insulin

Hyperlipidaemia (discussed with Consultants of biochemistry and metabolic medicine):
Commenced on Fibrate
Extremely low fat diet (10-15% of calorie intake) <20g/day



Results at 4 months

Cholesterol 3.9mmol (<5)
Triglycerides 1.8mmol (1.7)
HDL 1.1mmol (>1.2)
HbA1c 55mmol/mol (7.2%)

Conclusion

This case illustrates a young person whose initial presentation with eruptive xanthomas led to a diagnosis of new onset diabetes, complicated by severe hyperlipidaemia.

Hyperlipidaemia in children presenting in severe Diabetic Ketoacidosis (DKA) is a known, but rarely reported, phenomenon. The triglyceride levels in this case are higher than any other we could find. There is only one other report of a child presenting similarly with new onset diabetes and eruptive xanthomas that we could find[1].

In these cases care needs to be taken to evaluate the risk of pancreatitis, and there should be an awareness that lipaemia may make serum electrolyte measurement inaccurate – affecting DKA management[2].

Previous proposals to explain this phenomenon include genetic abnormalities of lipoprotein lipase[3], or a transient decrease in lipoprotein lipase activity secondary to insulin deficiency[2,4].

Management strategies are not standard and mostly derive from the treatment of hyperlipidaemic pancreatitis. These include plasmapheresis[2,5], fat-free total parenteral nutrition, insulin, heparin and antihyperlipidaemic drugs[5].

However, in many reports lipid profile improved with standard management of DKA alone.

There is no current work that we could find which attempts to explain the differences in severity of dyslipidaemia in children with new onset type 1 diabetes.

1 - Zabeen B, Khaled Z, Nahar J, Baki A, Amin F, Akhter S, Begum T, Azad K, Nahar N. *Hypertriglyceridemia associated with eruptive xanthomas and lipemia retinalis in newly diagnosed diabetes mellitus*. Mymensingh medical journal, July 2013, 22/3(591-595), 1022-4742
2 - Nyamugunduru G, Roper H. *A difficult case: Childhood onset insulin dependent diabetes presenting with severe hyperlipidaemia*. BMJ. Jan 4, 1997; 314(7073): 62–65.
3 - Karagianni C, Stabouli S, Roumeliotou K, et al. *Severe hypertriglyceridaemia in diabetic ketoacidosis: clinical and genetic study*. Diabet Med 2004;**21**:380–2.
4 - Abbate S, Brunzell J. *Pathophysiology of hyperlipidemia in diabetes mellitus*. Cardiovasc Pharmacol. 1990;16 Suppl 9:S1-7.
5 - Kyriakidis A, Raitsiou B, Sakagianni A, Harisopoulou V, Pyrgioti M, Panagopoulou A, Vasilakis N, Lambropoulos S. *Management of acute severe hyperlipidemic pancreatitis*. Digestion. 2006;73(4):259-64. Epub 2006 Aug 28.